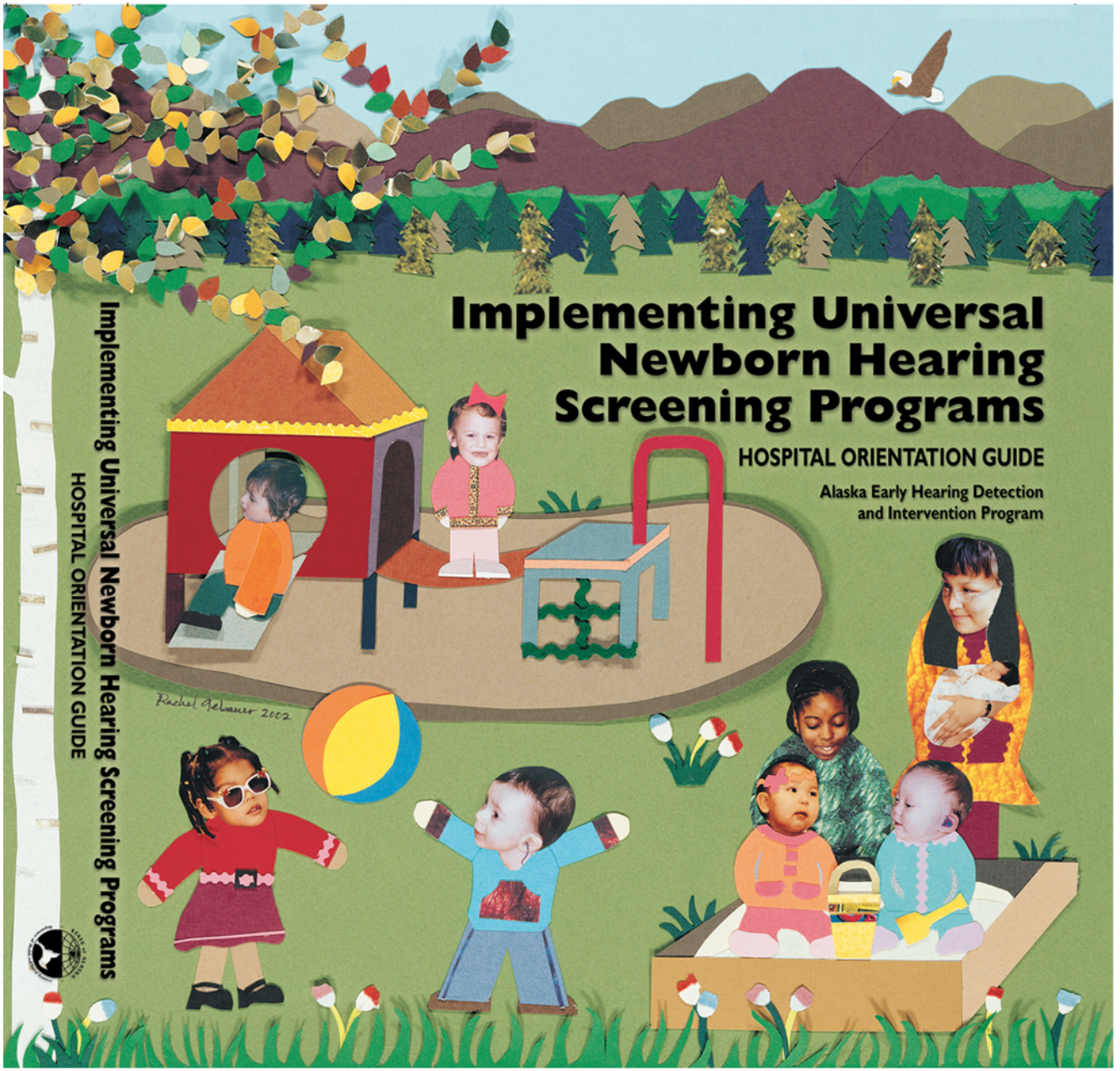
Implementing Universal Newborn Hearing Screening Programs
HOSPITAL ORIENTATION GUIDE
Alaska Early Hearing Detection and Intervention Program
Rachel Gehauer 2002
Implementing Universal Newborn Hearing Screening Programs
HOSPITAL ORIENTATION GUIDE

ACKNOWLEDGEMENTS

A special thanks to Lylis Olsen and the Ear Foundation of Arizona for their hard work in creating the Arizona 2001 Medical Guide. The Arizona Guide was used, with permission, as a template for the Alaska Early Hearing Detection & Intervention (EHDI) Provider Guide.

The Alaska EHDI Program also wishes to extend a deep appreciation to the following individuals for their time and expertise in the development and review of this guide.

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Major (S) Bridget McMullen, MA, CCC-A, US Air Force BSC
Lt. Col. Randall Ow, MD, US Air Force MC
Charlene Matesich, CCC-A
Jennifer Wingate, MD
Martin Beals, MD
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Table of Contents

BACKGROUND

The Alaska Early Hearing Detection and Intervention (AK EHDI) Program is a project of the State of Alaska, Department of Health and Social Services, Division of Public Health, Section of Maternal, Child and Family Health, Children's Health Unit. The program is funded by grants from the Health Resources Services Administration (HRSA) of the federal Maternal Child Health Bureau and the Center for Disease Control & Prevention (CDC). The goals of the program are to:

- Ensure that babies are screened for hearing loss at birth or shortly thereafter;
- Ensure that babies who refer from screening receive prompt and appropriate audiological and medical evaluation by three months of age;
- Ensure that babies who are identified as having a hearing loss and their families receive early intervention and other support services by six months of age; and
- Ensure that babies at risk for progressive or late onset hearing loss receive ongoing monitoring and evaluation.

A statewide advisory committee provides expertise and guidance to the program and assists in the development of guidelines, protocols, outreach materials, and evaluation activities. The AK EHDI Advisory Committee is comprised of pediatricians, parents, deaf and hard of hearing adults, early interventionists, public health nurses, audiologists, educators, speech-language pathologists, otolaryngologists, family practice physicians, public and private insurance representatives and others. Six taskforces were established by the committee, and focus on specific aspects of program development, including screening, diagnostics, intervention, media and outreach, data/evaluation, and family-centered care.

INTRODUCTION

The purpose of this document is to provide healthcare providers with guidelines for participation in the Alaska Early Hearing Detection and Intervention (AK EHDI) system. This guide is intended to be a

resource for healthcare providers and is not intended to replace clinical judgment for providing medical care.

The Alaska EHDI Healthcare Provider Guide is written to be consistent with the Joint Committee on Infant Hearing (JCIH) 2000 position statement (June 2000, *American Journal of Audiology* 9, 9-29) and the American Academy of Pediatrics (AAP) Policy Statement (February 1999, *Pediatrics* 103: 2, 527-30).

WHY EARLY HEARING DETECTION & INTERVENTION?

- Hearing impairment is the most common congenital disability in the United States and often has no visual indicators.
- The most critical period for speech and language development is from birth to age three.
- Late diagnosis and intervention can result in lifelong delays in speech, language, cognitive, and socio-emotional development.
- The average age of identification of hearing impairment in the absence of universal newborn screening is two to three years of age.
- Early identification and intervention are likely to result in normal language and cognitive development.

Universal newborn hearing screening is the first step in the early hearing detection and intervention process. Audiological and medical assessment, and linkage to early intervention services are critical subsequent steps.

Healthcare providers can expect families to look to them for answers and direction at each step of the EHDI process from the initial screening soon after birth to early intervention. Providers can assist by emphasizing the importance of the process and encouraging families to follow each of the steps below.

ROLE OF THE MEDICAL PROVIDER A STANDARD OF CARE

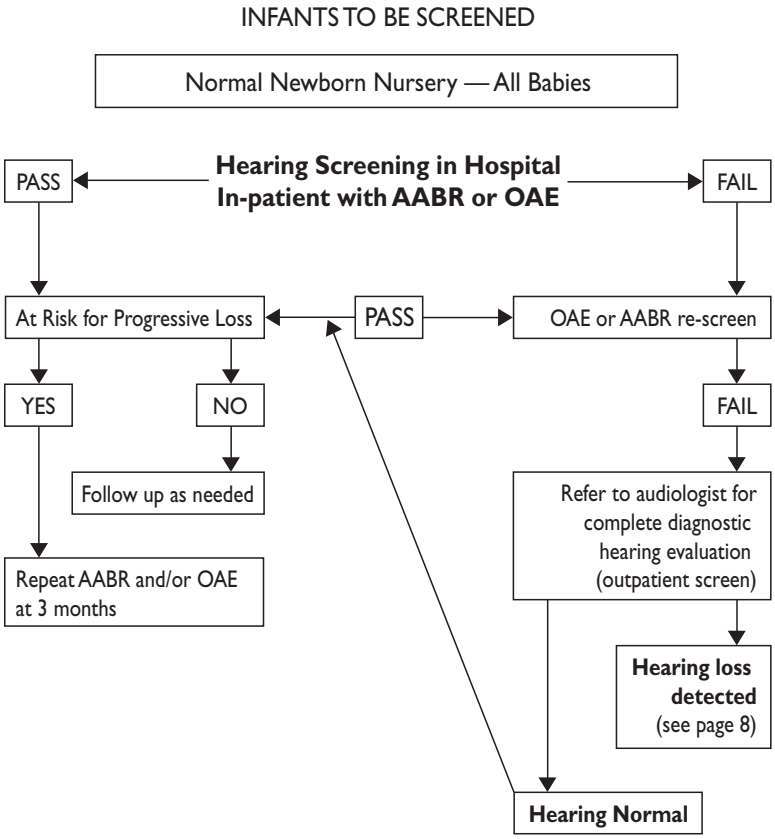
American Academy of Pediatrics 1999 Position Statement:

"Appropriate and necessary care for the infant with significant hearing loss should be directed and coordinated by the child's physician within the medical home, with support from appropriate ancillary services."

Healthcare providers can expect families to look to them for answers and direction at each stage of the EHDI process from the initial screening soon after birth to early intervention. Providers can assist by emphasizing the importance of the process and encouraging families to follow each of the steps below.

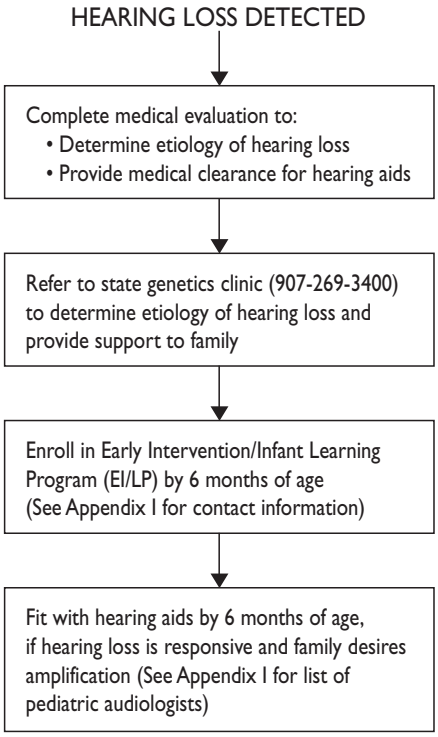
- Initial screen(s) at birthing hospital (or by 1 month of age).
- If initial screening(s) was missed at the birthing hospital, arrangements should be made at the nearest facility to perform screen.
- If initial screening(s) resulted in a "refer" (no pass), referral to a pediatric audiologist for a second screen.
- If second screening by an audiologist results in a refer, refer for complete frequency-specific Auditory Brainstem Response testing by a qualified audiologist by 3 months of age.

NEWBORN HEARING SCREENING PROTOCOL



- For infants confirmed on diagnostic assessment to have hearing impairment, the following protocol should be followed:
    1. Complete medical evaluation to explore etiology of hearing loss and provide medical clearance for hearing aids as soon as possible.
    2. Referral to state genetics clinic to explore etiology of hearing loss and provide support to family.
    3. Referral and enrollment in Early Intervention/Infant Learning Program (EI/ILP) by 6 months of age.
    4. Fit with hearing aids by 6 months of age, if hearing loss is responsive and family desires amplification.

NEWBORN HEARING SCREENING PROTOCOL

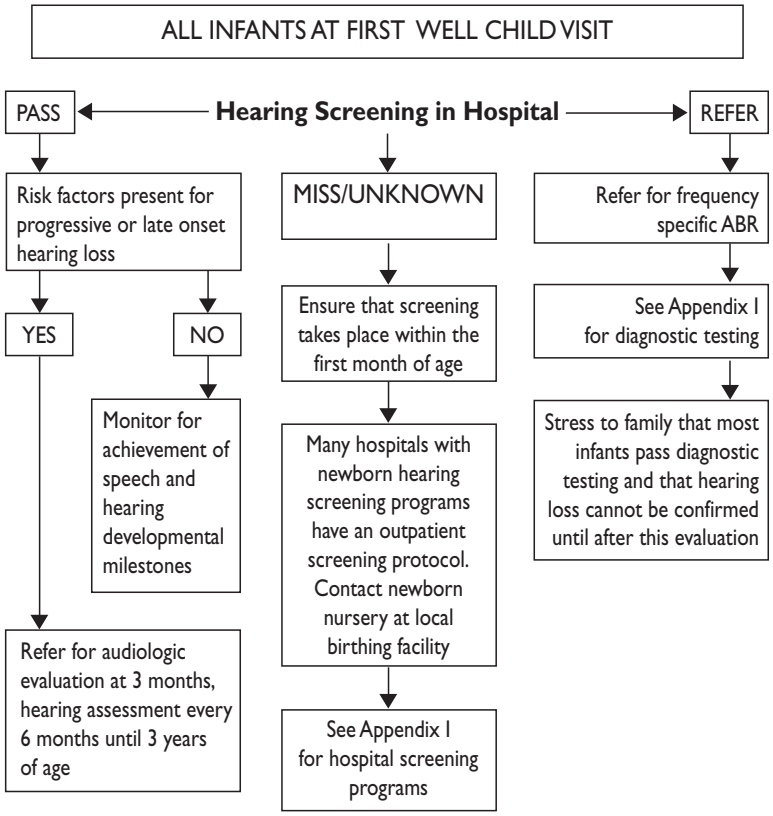


Screen for Hearing Loss by 1 Month

At the First Well Child Visit

Check that the infant was screened for hearing loss at her/his birth facility. If the infant was missed or did not pass, you should receive notification from the hospital (if you were the physician on record).

HEARING SCREENING
FIRST WELL CHILD VISIT PROTOCOL



Indicators for Children Who Are At Risk for Late Onset or Progressive Hearing Loss

The Joint Committee on Infant Hearing 2000 position statement suggests that the following indicators "…place an infant at risk for progressive or delayed-onset sensorineural and/or conductive hearing loss. Any infant with these risk indicators for progressive or delayed-onset hearing loss who has passed the birth screen should, nonetheless, receive audiologic monitoring every 6 months until age 3 years."

- Parental or caregiver concern regarding hearing, speech, language, and/or developmental delay.
- Family history of permanent childhood hearing loss.
- Postnatal infections associated with sensorineural hearing loss including bacterial meningitis.
- Head trauma.
- Recurrent or persistent otitis media with effusion for at least 3 months.
- Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or eustachian tube dysfunction.
- In-utero infections such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis.
- Neonatal indicators-specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation (ECHMO).
- Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and Usher's Syndrome.
- Neurodegenerative disorders, such as Hunter Syndrome, or sensory motor neuropathies, such as Friedreichs's ataxia and Charcot-Marie-Tooth Syndrome.

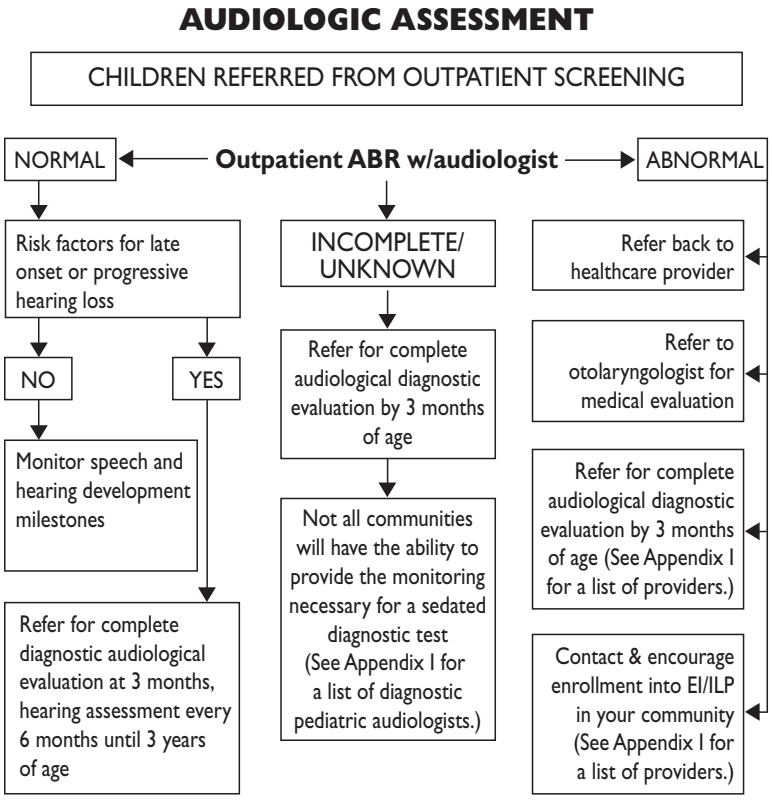
The incidence of congenital hearing loss is approximately:

- 3:1000 (0.3%) for all well babies
- 3:100 (3%) for Neonatal Intensive Care Unit (NICU) graduates

(National Center for Hearing Assessment & Management)

Complete Audiologic Assessment by 3 Months

For children who have referred from the outpatient screening:



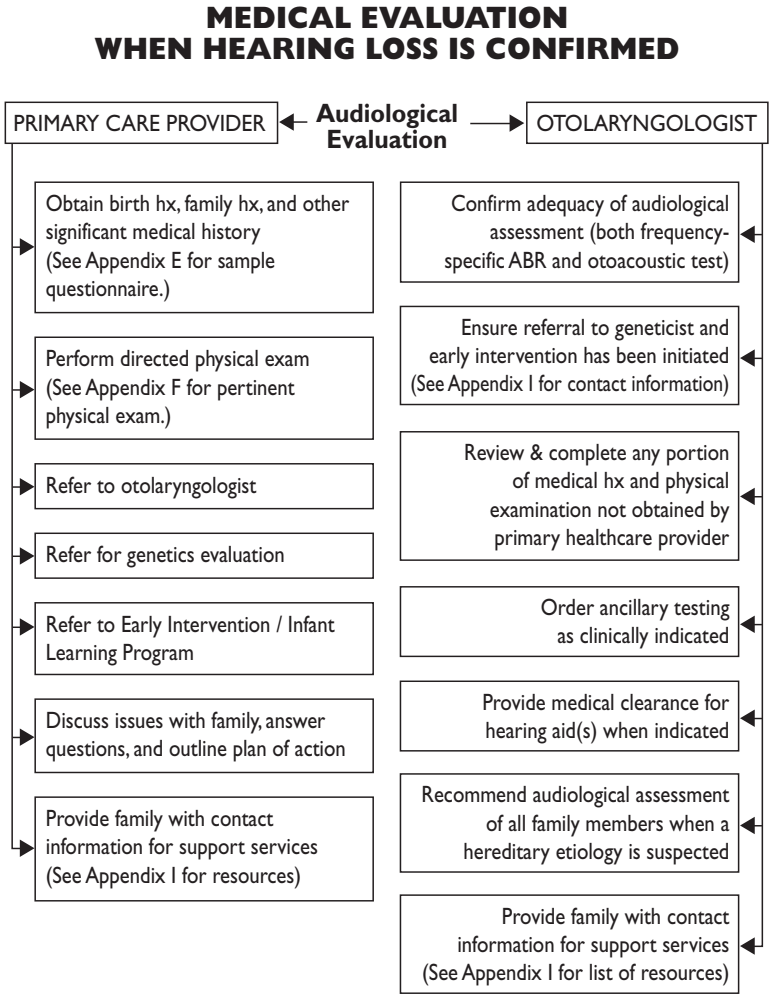
High Risk Factors Registry:

1. Family History
2. Defects of head & neck
3. Syndromal abnormalities
4. Low birthweight
5. Low Apgar scores
6. Hyperbilirubinemia
7. Anoxia
8. Bacterial and viral disease during pregnancy (i.e. rubella, chicken pox, meningoenciphalitis)
9. Congenital syphilis
10. Rh Incompatibility
11. Persistent fetal circulation
12. Ototoxic medication/drugs administered during pregnancy (i.e. neomycin, kanamycin)

Medical Evaluation When a Hearing Loss is Confirmed

When a Hearing Loss is Identified

The child's primary healthcare provider in concert with an otolaryngologist should evaluate the child for an etiology of hearing loss.



Component of a Family-Centered Plan of Care

Each provider should appreciate the potentially devastating effect(s) the diagnosis of hearing loss can have on a family. Communication should reflect both compassion and patience.

The evaluation of hearing impairment is complex and many providers do not frequently see children with hearing impairment. Emphasize to families that answers may not come quickly, and that patience and continued follow-up with different specialists is required to better serve their child.

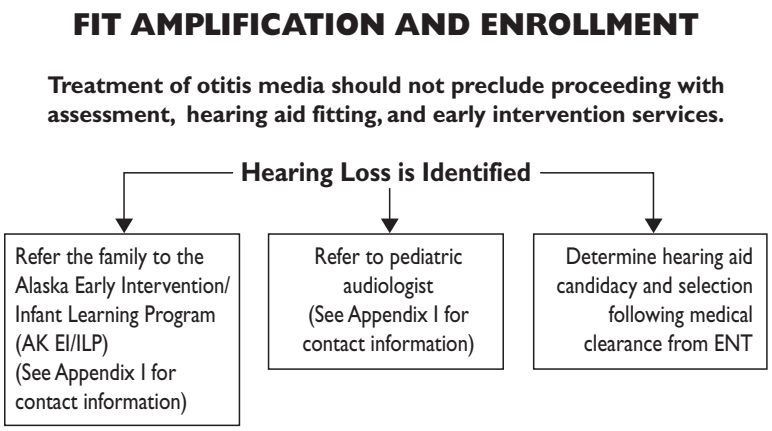
Some providers may prefer to schedule follow-up outpatient visits to complete their portion of the work-up. Incorporating an initial work-up for hearing loss during a well-baby check-up may not be feasible in some practices. The use of questionnaires (see Appendix E) can help facilitate the historical issues that need to be addressed. This preserves more time for the physical exam and time for questions and answers with the family.

The well-informed provider will be seen by the family as the child's strongest advocate. Discuss the role of the genetics evaluation, ENT evaluation, and EI/ILP as the treatment plan is developed. Provide specific support services and groups for families with hearing-impaired children. AK EHDI can assist offices that do not readily have the information. (See Appendix I for a list of resources available to families.)

Fit Amplification & Enroll in Early Intervention by 6 Months

When a Hearing Loss is Identified

Check that the family has enrolled in Early Intervention/Infant Learning Programs (EI/ILP) services, and is able to access resources for fitting hearing aids and specialty medical services.



Resources for Early Intervention

Alaska Early Intervention/Infant Learning Programs

In Alaska, the Early Intervention/Infant Learning Program (EI/ILP) is in place to provide appropriate services to families and their children (ages birth to three years of age) with developmental disabilities and delays. The program is available statewide and, therefore, every region of the state is served by one of 19 grantees. In addition, low incidence disorders like hearing impaired/deaf and deaf/blind dual sensory impairments have itinerant programs that team with the local programs to provide appropriate services.

Early intervention services include the following:

- Screening and Assessment
- Case Management
- Individualized Family Service Plan Development
- Transition

In addition, intervention services include home-based services from early childhood special educators, speech-language pathologists, occupational therapists, physical therapists, and others.

The purpose of these services is to help children and their families develop necessary skills through parent training and education, as well as, specialized intervention in therapy arenas.

The Alaska Early Intervention Hearing Resource (AEIHR) is a statewide itinerant program that provides specialized support and training for EI/ILP programs and families of children who experience a hearing impairment/deafness. The AEIHR is staffed with skilled Deaf educators who are familiar with the special developmental issues and educational challenges with children who experience a hearing impairment or deafness. (See Appendix I for list of EI/ILP providers.)

More about Early Intervention/ Infant Learning Program (EI/ILP), Hearing Aids and Cochlear Implants

Early Intervention (EI/ILP)

It is usually traumatic for a family to find out that their infant has a hearing loss. Parents will need assistance to work through grief issues and to answer the many questions that they will have. Early Intervention is critical to optimal outcomes and should be initiated as soon as the hearing loss is identified. Early Intervention, including family support and information, can begin even before the hearing aid(s) is fit and often before assessment is complete. (See Appendix I for EI/ILP providers.)

Hearing Aids

Clearance for Hearing Aids

State law and the Alaska Guidelines for Pediatric Audiological Assessment and Intervention require that medical clearance be obtained before hearing aids are fit on any child. JCIH 2000 states that management of OME, however, should not delay the prompt fitting of amplification unless there are medical contraindications.

Loaner Hearing Aids

The initial hearing aid fitting for newborns often requires more flexibility and options than may be needed later. The Alaska Assistive Device Loaner Program provides six month loaner advanced technology hearing aids for any child identified through newborn hearing screening who cannot secure quality devices through insurance. This allows the audiologist the time and flexibility to complete the assessment and to modify according to the changing needs of the child. The program can be accessed through any of the Audiologists with pediatric experience listed in Appendix I, or contact the State of Alaska, EHDI Program at (907) 269-3400.

Cochlear Implants

Nearly all children who are Deaf or Hard of Hearing, initially receive amplification through personal hearing aids. Some children who are Deaf, and who do not benefit from the use of hearing aids, may be candidates for cochlear implants. Evaluation for cochlear implantation can begin as soon as the hearing loss is identified. Usually, implantation does not occur until approximately 12-18 months of age.

Cochlear implants require extensive rehabilitation and a team approach to assess candidacy. Although cochlear implant surgery can be performed in Alaska, the extensive rehabilitation services and follow-up are not currently available in the state. Therefore, cochlear implant candidates must travel out of state for surgery.

Appendix A

REFERENCES

2000 Joint Committee on Infant Hearing Position Statement
August 2000, Audiology Today, Special Issue

1999 American Academy of Pediatric Policy Statement
February 1999, Pediatrics Vol. 103 No. 2 pp. 527-530.

1999 Arizona Pediatric Audiology Guidelines
Arizona Department of Health's Never Too Young Program

2001 Arizona Hospital Screening Guidelines
Available through the EAR Foundation of Arizona

SELECTED TEXTS

General Overview

Infants and Hearing
Deborah Hayes, PhD and Jerry Northern, PhD
1996, Singular Publishing Group, Inc.

Hearing Care for Children
Edited by Frederick N. Martin and John Greer Clark
1996, Allyn and Bacon

Hearing Aids and Habilitation

Amplification for Children with Auditory Deficits
Edited by Fred H. Bess, Judith S. Gravel
and Anne Marie Tharpe
1996, Bill Wilkerson Center Press

Genetics

Genetics and Hearing Impairment
Alessandro Martini, Andrew Read and Dafydd Stephens
1996, Singular Publishing Group, Inc.

American Journal of Medical Genetics
Seminars in Medical Genetics-*Hereditary Deafness*
Guest Editor William J Kimberling
September 24, 1999, Volume 89 Number 3

WEBSITES

www.infanthearing.org

www.lhh.org/parents/index.htm

www.hearingexchange.com

http://deafness.about.com/mbody.htm

Appendix B

COMMONLY USED ACRONYMS

AAP	American Academy of Pediatrics
ABR	Auditory Brainstem Response, also known as BAER and BSER
ALGO	Automated screening ABR equipment
BAER	Brainstem Auditory Evoked Response (ABR)
BSER	Brainstem Evoked Response (ABR)
CHIC	Center for Hearing Impaired Children
CI	Cochlear Implant
DPOAE	Distortion Product Otoacoustic Emissions
EHDI	Early Hearing Detection & Intervention
EI/ILP	Early Intervention / Infant Learning Program
ENT	Ear, Nose, and Throat Doctor
HA	Hearing Aid
JCIH	Joint Committee on Infant Hearing
OAE	Otoacoustic Emissions
OCSHCN	Office for Children with Special Health Care Needs
TEOAE	Transient Evoked Otoacoustic Emissions
UNHS	Universal Newborn Hearing Screening

APPENDIX C

INFORMATION ABOUT HEARING SCREENING IN NEWBORNS

Methods Used to Screen Newborns

Trained personnel can do either of the following screening methods in approximately 5 to 15 minutes, any time after birth. Screenings done before 24 hours of age are more likely to be affected by the presence of vernix/debris in the ear canal. Screenings done later in infancy are more likely to be affected by the presence of middle ear effusion. Neither of the screening methods directly measure the brain's ability to process sounds or true "hearing." Screening results only reflect the likelihood of hearing loss at the time of the screening. Some children are at risk for progressive or late onset hearing loss. (See Appendix H for high risk factors for hearing loss.)

1. ***Screening Auditory Brainstem Response***

   In an Auditory Brainstem Response (ABR) test, clicks are introduced into the ear canal from either a probe or over-the-ear coupler. An electroencephalographic (EEG) response is measured from electrodes placed on the scalp. The waveforms generated are measured against an internal template to determine a pass or refer result.

   ALGO© refers to a common brand of equipment used to perform the screening and is limited to screening infants under six months of age. Frequency-specific ABR used in assessment requires separate equipment and audiologists with pediatric experience.

2. ***Otoacoustic Emissions***

   In an otoacoustic emissions (OAE) test, a microphone placed in the ear canal measures sound waves generated in the cochlea in response to clicks or tone bursts. A pass or refer result is automatically generated based on preset criteria.

CONDITIONS REQUIRING COMBINED TESTING:

Auditory Neuropathy

Recently, auditory neuropathy was identified as affecting a small number of children with hearing loss, most of whom appear to be NICU graduates, some with hyperbilirubinemia. The exact prevalence of this disorder is unknown. This disorder is *only* identifiable through the combined use of ABR and OAE tests.

The primary characteristics of this disorder are absent, or abnormal ABR results and normal OAE results, which suggests that outer hair cells are functioning and that there is a problem with neural conduction.

Some NICU screening programs choose to use only ABR or a combination of OAE and ABR to detect this disorder. Most well baby screening programs target only significant sensorineural hearing loss for identification and therefore may use ABR or OAE.

Appendix D

INFORMATION ABOUT HEARING ASSESSMENT IN INFANTS

Methods Used to Assess & Diagnose Hearing Loss in Infants & Toddlers

1. ***Auditory Brainstem Response (ABR) Testing***
   An ABR test is the accepted method of evaluating infants under the developmental age of 6 months. An ABR test allows not only an estimation of hearing thresholds, but can also indicate the possibility of a conductive component to the hearing loss and can give some frequency specific information. Frequency-specific ABR is important when assessing hearing for determining hearing aid fitting.
2. ***Behavioral Assessment***
   Behavioral testing should also be done to validate the physiological results obtained. Behavioral test results become more accurate as a baby approaches 6 months of age.
3. ***Tympanometry***
   Due to the plasticity of the newborn's ear canal, normal tympanometry results cannot be considered accurate until approximately 6 months of age. High frequency tympanometry is more accurate but not readily available in all sites and is rarely available outside otolaryngology and audiology offices.

Unilateral Hearing Loss

Children with a unilateral hearing loss should receive the same medical assessment as children with bilateral hearing loss to determine etiology and monitor loss. The Alaska Early Intervention/Infant Learning Program does serve children with unilateral hearing loss.

Hereditary Hearing Loss

Nearly 200 (JCIH-2000) syndromic and nonsyndromic forms of hearing loss have been identified. Most congenital hearing loss is hereditary. For many of these children the associated clinical findings can be of importance in patient management. Related medical specialty ar-

eas may include: developmental pediatrics, neurology, ophthalmology, cardiology, and nephrology. (Contact State of Alaska, Genetics Clinic, (907) 269-3400 for further information.)

Insurance Coverage for Assessment

Some health plans require pre-authorization from the primary healthcare provider before the ABR is completed. Unfortunately, this can delay services beyond the time when the ABR can be performed unsedated and may result in the need for authorization for a sedated ABR. This may also be complicated by the need for referral to a pediatric audiology center capable of performing a frequency-specific ABR, which may be outside the preferred provider list for the health plan.

It is in the best interest of the child and family to expedite the process as much as possible to meet the goals of early detection and intervention. Delayed identification beyond six months of age is likely to result in delayed speech and language acquisition, as well as other preventable outcomes.

An ABR test is also covered by Denali KidCare, Indian Health Services, and most private health plans.

Genetics Clinics

The State of Alaska does not have a medical school or genetics laboratory. Services are available through the Alaska Genetics Clinics that offer information about genetics disorders and genetic evaluations for diagnosis. The clinic staff includes clinical geneticists from the University of Washington, Children's Hospital and Regional Medical Center in Seattle, a genetic counselor from the State of Alaska, Section of Maternal, Child & Family Health, and local public health nurses. The process includes working with families for pre-clinic preparation and post-clinic follow-up, as well as, further testing if necessary.

Appendix E

FAMILY QUESTIONNAIRE	Yes	No	Don't know
PARENT CONCERNS			
Do you have concerns about your child's ability to hear voices or sounds?			
Is there a history of deafness or hearing loss in children in your family?			
Has anyone in your family suffered an unexplained and sudden death or seizures?			
PRENATAL HISTORY			
Was the pregnancy complicated by maternal infections? (CMV, Rubella, HIV, Herpes, Syphilis, Toxoplasmosis)			
Did the mother take any prescribed medication during pregnancy?			
Did the mother take any over-the-counter medications or herbal supplements during pregnancy?			
Did the mother use any other drugs during her pregnancy? (Cocaine, marijuana, etc.)			
Did the mother receive regular prenatal care?			
Did the mother receive any immunizations during pregnancy?			
Does the mother have any serious medical conditions?			
Does the mother have a history of miscarriages?			
Did the mother care for a cat during her pregnancy?			

FAMILY QUESTIONNAIRE	Yes	No	Don't know
BIRTH HISTORY			
Was the baby born full-term?			
The baby weighed _____lbs, ____oz at birth.			
Was the baby in any distress during delivery? (APGAR <4)			
Was there Rh incompatibility?			
Did the baby have jaundice requiring treatment in the hospital?			
Was the baby admitted to the intensive care unit of the hospital?			
Did the baby experience meconium aspiration at birth?			
Did the baby need IV antibiotics after birth?			
MEDICAL HISTORY			
Has the baby had any head injury or significant trauma since birth?			

Appendix E *continued*

FAMILY QUESTIONNAIRE	Yes	No	Don't know
BIRTH HISTORY			
Term SVD unremarkable			
Antenatal infections			
Pre-term/post-term			
Delivery SVD/c-section: any signs of fetal distress?			
meconium aspiration?			
Rh incompatibility			
Neonatal ICU admission			
Sepsis or infection requiring IV antibiotic therapy (See Appendix H for listing of ototoxic drugs)			
MATERNAL HISTORY			
Healthy			
History of recurrent spontaneous abortions			
Adequacy of prenatal care			
Prescribed drug use during pregnancy			
OTC drug use during pregnancy			
Illicit drug use during pregnancy			
ETOH use during pregnancy			
Immunizations during / around pregnancy			
Rubella status			

FAMILY QUESTIONNAIRE	Yes	No	Don't know
MATERNAL HISTORY continued			
Infections during pregnancy			
Toxo/rubella/CMV/herpes/syphilis			
STD history			
Cat exposure (i.e., care of cat)			
Other serious medical conditions			
FAMILY HISTORY			
Known hereditary hearing loss			
Deaf or blind relatives			
Family member with hearing aid before the age of 40			
Any congenital birth defects			
Any relative with history of unexplained sudden death or unexplained recurrent syncope			

Appendix F

PHYSICAL EXAM

The physical examination of the infant referred after confirmation of significant hearing loss has two primary goals:

1. Identify possible cause of hearing loss
   a. Occlusion of external ear canal with foreign material, vernix, or cerumen
   b. Atresia of the external auditory canal
   c. Fluid in the middle ear space

2. Identify presence or absence of stigmata known to be associated with syndromic forms of hearing loss. These stigmata usually involve craniofacial malformations, ocular changes, ear malformations, neck masses, integumentary changes, and malformations of the digits
   a. Face
      i. Hemifacial microsomia -(Goldenhar's)
      ii. Mandibulo - (Treacher-Collins)
      iii. (Crouzon's)
   b. Neck
      i. Lateral neck masses or cervical pits - (Melnick-Fraser or branchio-oto-renal syndrome)
      ii. Goiter - (Pendred)
   c. Eyes
      i. Coloboma
      ii. Iris color - Heterochromia (Waardenburg)
      iii. Eyebrow formation – (Waardenburg)
      iv. Retinitis Pigmentosa – (Usher)
      v. Severe Myopia – (Stickler, Goldenhar's)
   d. Ears
      i. Microtia
      ii. Canal atresia
      iii. Canal occlusion
      iv. Middle ear fluid

e. Limbs
    i. Syndactyly
    ii. Limb defects – progressive arthritis – (Stickler)
f. Skin
    i. White forelock – (Waardenburg)
g. Mouth
    i. Cleft palate
h. Neuro
    i Epilepsy
    ii Stroke-like episodes -(Mitochondrial defects)

Appendix F

HEARING AND SPEECH DEVELOPMENTAL MILESTONES CHECKLIST

The following are developmental guidelines for children with normal hearing.

0-3 months
- Jumps at a sudden, loud noise
- Calms down when you speak

3-6 months
- Turns head or moves eyes to find your voice
- Plays at making noises and sounds

6-10 months
- Reacts to hearing his/her name
- Begins to understand easy words like "no" and "bye-bye"

10-15 months
- Repeats simple words and sounds you make
- Uses two to three words other than "ma-ma" and "da-da"

18 months
- Follows simple spoken directions
- Uses seven or more true words

24 months
- Understands when you call from another room
- Points to body parts when asked

Appendix G

ANCILLARY TESTING

The decision to send a patient for radiologic or laboratory testing can be left for the otolaryngology consultant to decide. Cost-effective use of ancillary testing requires a specialist versed in the entire spectrum of hearing disorders. The specialist must consider the historical factors along with physical findings to make decisions regarding ancillary testing.

The following tests can yield important information in the search for an etiology of hearing loss. The conditions tested for can sometimes be rare, but potentially serious. Clinical suspicion may make many of the tests contraindicated.

Consultations

Genetics (hereditary hearing impairment, genetic testing)
Ophthalmology (screening for Retintis Pigmentosa)

Laboratory testing

Serology for CMV, toxo, rubella, syphilis, herpes
Thyroid function tests (Pendred's syndrome)
Urinalysis (Alport's syndrome)

Radiology testing

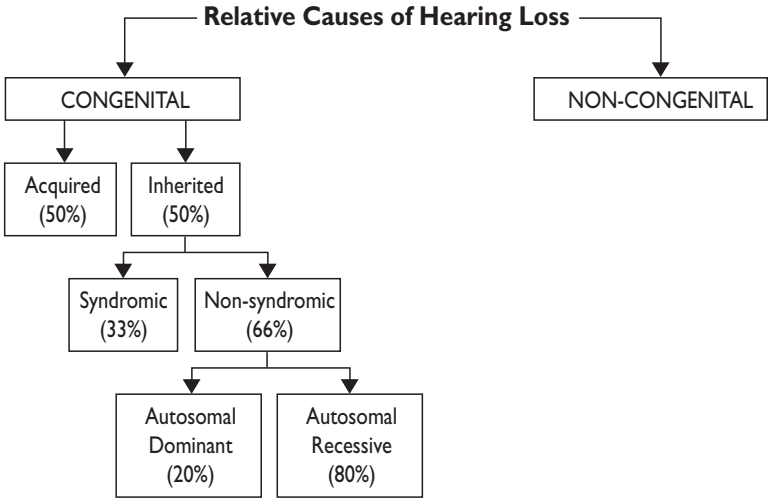
CT scan (Michel's, Mondini deformity, enlarged vestibular aqueduct)
MRI
Perchlorate discharge test (Pendred's syndrome)
Renal ultrasound (branchial-oto-renal syndrome)

Other

EKG (Jervel and Lange-Nielsen)

Appendix H

RELATIVE CAUSES OF HEARING LOSS



High Risk Factors Registry:

1. Family History
2. Defects of head & neck
3. Syndromal abnormalities
4. Low birthweight
5. Low Apgar scores
6. Hyperbilirubinemia
7. Anoxia
8. Bacterial and viral disease during pregnancy (i.e. rubella, chicken pox, meningoenciphalitis)
9. Congenital syphilis
10. Rh Incompatibility
11. Persistent fetal circulation
12. Ototoxic medication/drugs administered during pregnancy (i.e. neomycin, kanamycin)

Appendix H

HIGH RISK FACTORS FOR HEARING LOSS

1. Parental concern of hearing impairment

2. NICU admission for greater than 48 hours

3. Stigmata or other physical findings suggestive of syndromic hearing loss

4. Family history of permanent sensorineural childhood hearing loss

5. Craniofacial anomalies, especially those associated with malformations of the pinna or ear canal

6. In utero infections such as CMV, herpes, rubella or toxoplasmosis

7. Post-natal infections such as sepsis or bacterial meningitis

8. Low birthweight

9. • Respiratory abnormalities
   • Respiratory distress syndrome
   • Bronchopulmonary dysplasia
   • Prolonged mechanical ventilation

10. Hyperbilirubinemia

11. Head trauma

12. Recurrent or persistent otitis media with effusion greater than 3 months

13. Low Apgar scores

Appendix H

OTOTOXIC MEDICATIONS

Aminoglycosides
- **Gentamicin**
- **Kanamycin**

Vancomycin
Furosemide
Streptomycin
Dihydrostreptomycin
Neomycin
Polymyxin B
Erythromycin (high doses in treatment of legionella)

Carbon monoxide
Mercury
Gold
Lead
Arsenic
Aniline dyes

Ethacrynic acid
Cisplatin
Chloroquine
Quinine
Salicylates
Polybrene
Nitrogen mustard
Thalidomide

Trauma

Kernicterus (hyperbilirubinemia) greater than 340 mmol/L is harmful to CNS development – can lead to cerebral palsy, seizures

Appendix H

INHERITED DISORDERS

Autosomal dominant

- Branchiootorenal Syndrome (Melnick-Fraser)
- DFNA (connexin 26) 1-15
- Crouzon's Syndrome
- Neurofibromatosis Type II
- Osteogenesis Imperfecta
- Stickler's Syndrome
- Treacher-Collins Syndrome
- Waardenburg's Syndrome

Autosomal recessive

- DNFB (connexin 26) 1-19
- Alexander Aplasia
- Alport's Syndrome
- Insley's Syndrome
- Jervell and Lange-Nielsen
- Pendred's Syndrome (associated with enlarged vestibular aqueduct)
- Mondini Deformity
- Michel Deformity
- Schiebe Aplasia
- Usher's Syndrome

X-linked

- Albinism Deafness Syndrome
- DFN 1-6
- Norrie's Disease
- X-linked mixed hearing loss with Stapes Gusher

Mitochondrial

Unknown

Enlarged vestibular aqueduct

Sporadic (not-inherited)

- Goldenhar's

(Connexin 26 is a gene that codes for gap junction proteins in the outer hair cells of the cochlea. Over 200 mutations of the gene are known, usually point mutations. Depending on the mutation, the protein coded for can be partially functional explaining the varied expression and degrees of hearing loss seen in affected individuals)

Appendix H

ACQUIRED CAUSES OF HEARING LOSS

Infections

Congenital:

- Viral
  - Cytomegalovirus
  - Rubella
  - Herpes simplex
- Toxoplamosis
- Syphilis

Neonatal:

- Sepsis
- Mumps
- Measles
- Bacterial meningitis

Appendix I

ALASKA UNIVERSAL NEWBORN HEARING SCREENING PROGRAMS

It is likely that phone numbers will change. If a more current number is needed, contact the hospital in your community.

Anchorage
Alaska Native Medical Center ..... (907) 729-1422
Alaska Regional Hospital ..... (907) 264-1331
Providence Alaska Medical Center ..... (907) 562-2211
Elmendorf Air Force Base Hospital ..... (907) 580-1663

Barrow
Samuel Simmonds Hospital ..... (907) 852-9331
(not screening as of 3/03)

Bethel
Yukon Kuskokwim Health Center ..... (907) 543-6473

Cordova
Cordova Public Health Center ..... (907) 424-4547
(expected to begin screening by 6/03)

Dillingham
Kanakanak Hospital ..... (800) 478-5201

Fairbanks
Bassett Army Hospital ..... (907) 353-5158
Fairbanks Memorial Hospital ..... (907) 458-5227

Homer
South Peninsula Hospital ..... (907) 235-0256
(not screening as of 3/03)

Juneau
Bartlett Memorial Hospital ..... (907) 463-8657
(not screening as of 3/03)

Ketchikan
Ketchikan General Hospital ..... (907) 228-8300

Kotzebue
Maniilaq Hospital ..... (907) 442-7304

Kodiak
Providence Kodiak Island Medical Center ..... (907) 486-3281

Matsu Valley
Valley Hospital ..... (907) 746-8600

Nome
Norton Sound Regional Hospital ..... (907) 443-3297

Petersburg
Petersburg Community Hospital……. ..... .(907) 772-4291
Petersburg Public Health Center……. ..... (907) 772-4611
(expected to begin screening by 6/03)

Sitka
Sitka Community Hospital…… ..... .(907) 747-1722
Sitka Public Health Center……. ..... (907) 747-3255
(expected to begin screening by 6/03)

Mt. Edgecumbe Hospital (SEARHC) ..... (907) 966-8379

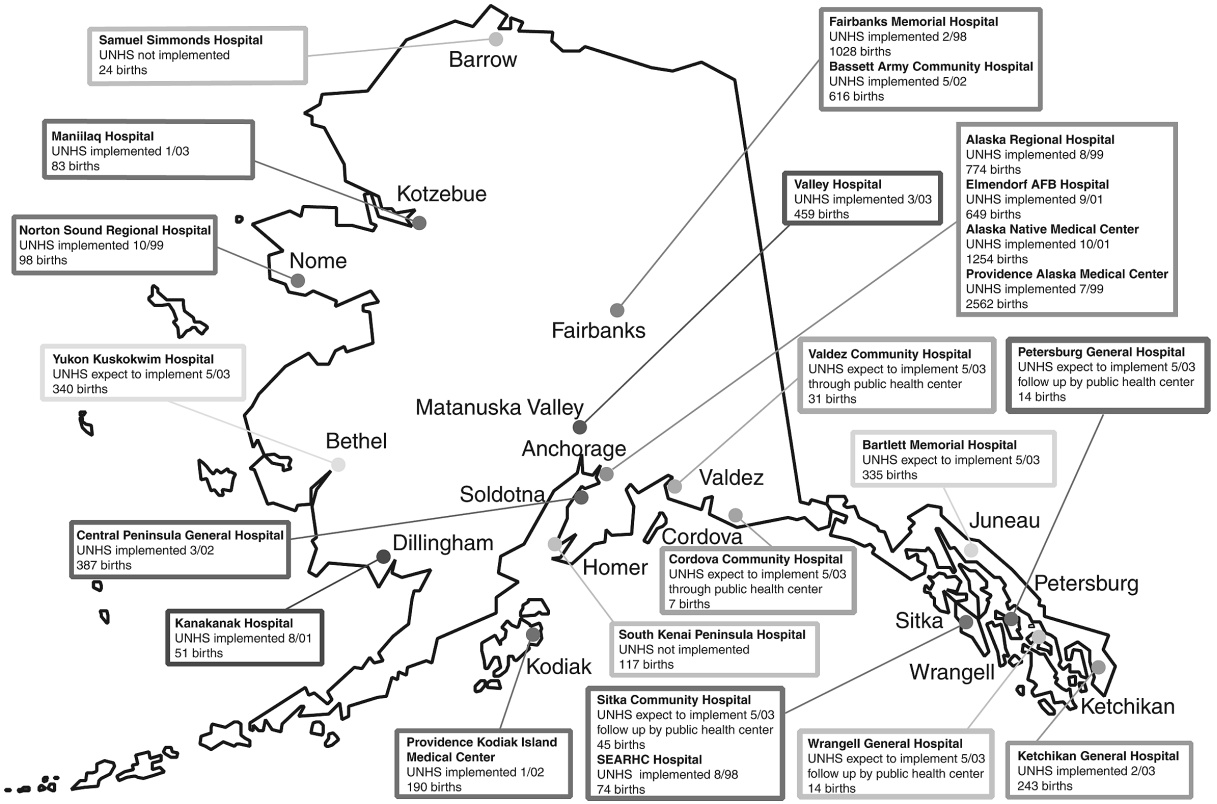
Soldotna
Central Peninsula Hospital ..... (907) 262-8107

Valdez
Valdez Public Health Center ..... (907) 835-4612
(expected to begin screening by 6/03)

Wrangell
Wrangell General Hospital ..... (907) 874-7000
Wrangell Public Health Center ..... (907) 874-3615
(expected to begin screening by 6/03)

Locations of Newborn Hearing Screening Hospitals

2002 births



Appendix I

AUDIOLOGIC DIAGNOSTIC EVALUATION PROVIDERS IN ALASKA

✱ Non-sedated diagnostic evaluations ▲ Sedated diagnostic evaluations

ANCHORAGE

▲ **Alaska Native Medical Center**
Linda Erb, David Brueggemann
Pediatric Audiologists
4315 Diplomacy Drive, Anchorage, AK 99508
(907) 729-1422

▲ **Alaska Regional Hospital**
Carol Ford, Nancy Johnston
Electroneurodiagnostic Technologists
2801 Debarr Rd., EEG Department, 5th Floor
Anchorage, AK 99508
(907) 264-1510

Audiology Associates
Tom McCarty
Pediatric Audiologist
1200 Airport Heights, Anchorage, AK 99508
(907) 278-6400

▲ **Headquarters 3rd Medical**
Capt. Bridget McMullen
Pediatric Audiologist
24800 Hospital Drive, Elmendorf AFB, AK 99506-3700
(907) 580-5804

Northern Hearing Services, Inc.
Joyce Sexton, Jess Buri, Karen Pastell, Alyson Kantorowski
Pediatric Audiologists
4200 Lake Otis Parkway, Ste. 302, Anchorage, AK 99508
(907) 561-1326

OVIDERS

evaluations

▲ **Providence Hospital Audiology**
Charlene Matesich
Pediatric Audiologist
3300 Providence Drive, Anchorage, AK 99519
(907) 261-3650

BETHEL

✸ **Yukon Kuskokwim Delta Hospital**
Mike Comerford
Pediatric Audiologist
PO Box 287, Bethel, AK 99559
(907) 543-6473

DILLINGHAM

✸ **Bristol Bay Native Corporation**
Deborah Burton
Pediatric Audiologist
PO Box 130, Dillingham, AK 99508
(907) 478-5201

FAIRBANKS

Bassett Army Hospital
Capt. Melissa Leccese
100 Gaffney Road, #7400, Fort Wainwright, AK 99703
(907) 353-5212

✸ **Northern Audiology & Hearing, ENT Clinic**
Art Larson
Pediatric Audiologist
1919 Lathrop, Suite 207, Fairbanks, AK 99701-5995
(907) 456-7768

HOMER

Susan Bunting
Pediatric Audiologist
PO Box 771, Homer, AK 99603
(907) 235-2381

JUNEAU

* **Northland Audiology**
Alver Rongstad
Pediatric Audiologist
PO Box 32257, Juneau, AK 99803
(907) 789-6780

KENAI PENINSULA

* **Peninsula Hearing Services**
Karen Martin
Pediatric Audiologist
167 Warehouse Avenue, Soldotna, AK 99669
(907) 262-3224

NOME/NORTON SOUND

* **Norton Sound Regional Hospital**
Dan Knudsen, Phil Hofstetter
Pediatric Audiologists
PO Box 966, Nome, AK 99762
(907) 443-3297

SITKA

SEARHC/Mt. Edgecumbe Hospital
Kala Stone
Pediatric Audiologist
222 Tongass Drive, Sitka, AK 99835
(907) 966-8379

Providers were identified through a survey. Every effort was made to provide accurate information, however, phone numbers and provider information may change. The Alaska Early Hearing Detection & Intervention Program does not specifically endorse or recommend any providers.

Appendix I

ALASKA EI/ILP PROGRAMS IN YOUR COMMUNITY

ANCHORAGE
Programs for Infants & Children, Inc. (PIC)
561-8060

Alaska Early Intervention Hearing Resource (AEIHR)
742-4273

Vision Impairment Services For Infants & Toddlers (VISIT)
562-7372

BARROW
North Slope Borough School District Infant Learning Program
852-9676

BETHEL
Community Services Family Infant Toddler Program
543-3690

CHUGIAK
Family Outreach Center Understanding Special Needs, Inc. (FOCUS)
688-0282

COPPER CENTER
Copper River Basin Infant Learning Program
822-5655

CORDOVA
Prince William Sound Infant Learning Program
424-3425

DILLINGHAM
Bristol Bay Area Health Corporation Infant Learning Program
842-3398

FAIRBANKS
Alaska Center for Children & Adults, Inc.
456-4003

Tanana Chiefs Conference Infant Learning Program
452-8251

HOMER
Children's Services Birth 2 Three
235-6044

HAINES
REACH Infant Learning Program
766-2750

JUNEAU
REACH Infant Learning Program
586-8228

KETCHIKAN
Community Connections Early Learning Program
225-7825

KODIAK
KANA Kodiak Infant Learning Program
486-4643

KOTZEBUE
Northwest Arctic Borough School District
Early Learning and Family Program
442-3472

NOME
Norton Sound Health Corporation Infant Learning Program
443-3298

PETERSBURG
REACH Infant Learning Program
772-3488

SEWARD
SeaView Community Services
224-5257

SITKA
Center for Community Early Learning Program
747-6960

SOLDOTNA
Frontier Community Services Early Intervention
262-3144

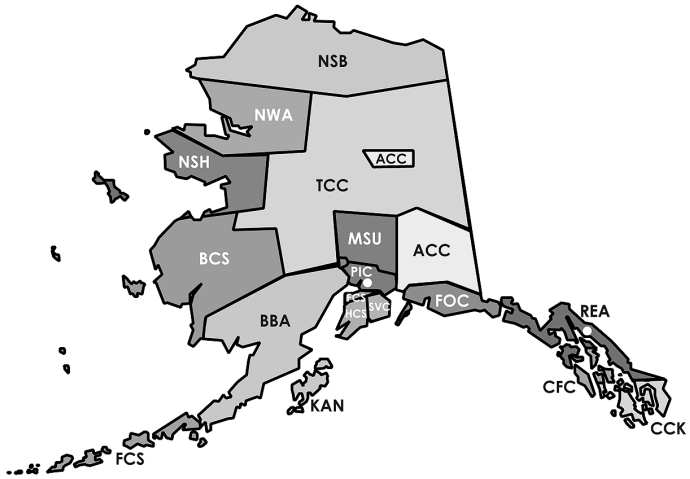
UNALASKA
Frontier Community Services Early Intervention
262-3144

VALDEZ
Infant Learning Program
835-4323

WASILLA
Mat-Su Services for Children & Adults Infant Learning Program
352-1200

STATE OF ALASKA
Dept. Health & Social Services
Early Intervention/Infant Learning Program
(907) 269-3400
http://www.hss.state.ak.us/dph/mcfh/

EI/ILP Providers, 2001



ACC ACCA/Copper River/Fairbanks/Valdez
BBA Bristol Bay Area Health Corp./ Dillingham
BCS Bethel Community Services/Bethel
CCK Community Connections/Ketchikan
CFC Center for Community/Sitka
FCS Frontier Community Services/Soldotna/ Aleutian/Pribilofs
FOC FOCUS, Inc./Chugiak/Cordova
HCS Homer Childrens' Services/Homer
KAN Kodiak Area Native Assn./Kodiak
MSU Mat-Su Services for Children & Adults, Inc./Wasilla
NSB North Slope Borough/Barrow
NSH Norton Sound Health Corp./Nome
NWA Northwest Arctic School District/Kotzebue
PIC Programs for Infants & Children/Anchorage/Whittier
REA REACH/Juneau/Haines/Petersburg
SVC SeaView Community Services/Seward
TCC Tanana Chiefs Conference/Fairbanks (Interior)

Note: Statewide consultant programs for vision and hearing are based in Anchorage.

Every effort was made to provide accurate information, however, phone numbers and provider information may change. The Alaska Early Hearing Detection & Intervention Program does not specifically endorse or recommend any providers.

Appendix I

PEDIATRIC AUDIOLOGISTS IN ALASKA

ANCHORAGE

Joyce Sexton, Jess Buri, Karen Pastell, Alyson Kantorowski
Northern Hearing Services, Inc.
4200 Lake Otis Parkway, Ste. 302, Anchorage, AK 99508
(907) 561-1326

Charlene Matesich
Providence Hospital Audiology
3300 Providence Drive, Anchorage, AK 99519
(907) 261-3650

Tom McCarty
Audiology Associates
1200 Airport Heights, Anchorage, AK 99508
(907) 278-6400

Linda Erb, David Brueggemann
Alaska Native Medical Center
4315 Diplomacy Drive, Anchorage, AK 99508
(907) 729-1422

Capt. Bridget McMullen
Headquarters 3rd Medical
24800 Hospital Drive, Elmendorf AFB, AK 99506-3700
(907) 580-5804

BETHEL

Mike Comerford
Yukon Kuskokwim Delta Hospital
PO Box 287, Bethel, AK 99559
(907) 543-6473

DILLINGHAM

Deborah Burton
Bristol Bay Native Corporation
PO Box 130, Dillingham, AK 99508
(907) 478-5201

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Art Larson
Northern Audiology & Hearing, ENT Clinic
1919 Lathrop, Suite 207, Fairbanks, AK 99701-5995
(907) 456-7768

Capt. Melissa Leccese
Bassett Army Hospital
100 Gaffney Road, #7400, Fort Wainwright, AK 99703
(907) 353-5212

HOMER
Susan Bunting
PO Box 771, Homer, AK 99603
(907) 235-2381

JUNEAU
Alver Rongstad
Northland Audiology
PO Box 32257, Juneau, AK 99803
(907) 789-6780

KENAI PENINSULA
Karen Martin
Peninsula Hearing Services
167 Warehouse Avenue, Soldotna, AK 99669
(907) 262-3224

NOME/NORTON SOUND
Dan Knudsen, Phil Hofstetter
Norton Sound Regional Hospital
PO Box 966, Nome, AK 99762
(907) 443-3297

SITKA
Kala Stone
SEARHC/Mt. Edgecumbe Hospital
222 Tongass Drive, Sitka, AK 99835
(907) 966-8379

Appendix I

ALASKA GENETICS CLINIC

The Genetics Clinic offer diagnostic and consultative services to medical professionals and health service agencies for Alaskan families with inherited disorders, chromosomal anomalies and birth defects. Medical genetic services are available to all persons in Alaska. Appointments are private and all information is strictly confidential. Medical geneticists, contracted through the Children's Hospital & Medical Center in Seattle, conduct clinics in six Alaska cities: Anchorage, Bethel, Fairbanks, Juneau, Ketchikan, and Sitka. Throughout the year a genetic counselor is on staff in Anchorage, and medical geneticists in Seattle are available for medical consultation.

For referrals/appointments, and additional information regarding medical genetic services, contact the local clinic coordinator.

Anchorage: Christy LeBlond, MS 269-3430
Bethel: Donna Chris, PHN 543-2110
Dillingham: Janis Hutsik, PHN 842-5981
Fairbanks: Dennette Marks, PHN 451-1665
Juneau: Victoria Gibson, PHN 465-1257
Ketchikan: Susan Medel, PHN 225-4350
Kodiak: Darsha Spalinger, PHN 486-3319
Sitka: Michelle Kennedy, PHN 747-3255

Directors - Genetic
Roberta Pagon, MD ..... (206) 526-2056
Ian Glass, MD ..... (206) 526-2056
Director - Metabolic
C. Ronald Scott, MD ..... (206) 543-3370

Appendix J

EDUCATIONAL RESOURCES

Websites

National Center for Hearing Assessment and Management (NCHAM)
Voice: (435) 797-3584
Email: mail@infanthearing.org
Internet: www.infanthearing.org
Description: The goal of the National Center for Hearing Assessment and Management (NCHAM - pronounced "en-cham") at Utah State University is to ensure that all infants (newborns) and toddlers with hearing loss are identified as early as possible and provided with timely and appropriate audiological, educational, and medical intervention. NCHAM receives funding from federal, state, and private sources to conduct research, develop training materials, provide training and technical assistance, and disseminate information about early identification and management of hearing loss.

Alexander Graham Bell Association for the Deaf and Hard of Hearing (A.G. Bell)
Voice: (202) 337-5220
TTY: (202) 337-5221
Toll-free: 1-800-HEAR-KID (1-800-432-7543)
Email: info@agbell.org
Internet: www.agbell.org
Description: A nonprofit, membership-based information center on hearing loss. Focuses specifically on children with hearing loss, providing ongoing support and advocacy for parents, professionals, and other interested parties. Information available to parents includes publications, funding sources, pamphlets, conferences, and scholarship program information.

American Society for Deaf Children (ASDC)
Voice: (717) 334-7922
TTY: (717) 334-7922
Toll-free: 1-800-942-ASDC (1-800-942-2732)
Email: ASDC1@aol.com
Internet: www.deafchildren.org
Description: A nonprofit organization designed to educate, empower and support parents and families of children who are deaf or hard-of-hearing. Helps families find meaningful communication options, particularly through the use of sign language, in their home, school, and community.

Boystown National Research Hospital
Voice: (402) 498-6511
TTY: (402) 498-6543
Toll-free: 1-800-282-6657
Email: moeller@boystown.org
Internet: www.boystownhospital.org
Description: A nonprofit hospital that is internationally recognized for research and treatment of childhood deafness and communication disorders. Programs include the Center for Audiology and Vestibular Services, the Center for Childhood Deafness, Language, and Learning, and the Center for Medical/Surgical Services. Produces videotapes designed to help families learn signs and read effectively with young children who are deaf.

For more information about services in Alaska contact:
Alaska Department of Health and Social Services
Division of Public Health
Section of Maternal, Child and Family Health
Alaska Early Hearing Detection & Intervention Program
http://www.hss.state.ak.us/dph/mcfh
(907) 269-3400

Alaska Universal Newborn Hearing Screening
Early Hearing Detection and Intervention Program

Educational Materials

In an effort to promote public awareness regarding universal newborn hearing screening, a four-pronged approach covering all steps involved throughout the diagnosis of a possible hearing loss, was developed by the Alaska Early Hearing Detection & Intervention Program. For copies of these materials, contact the EHDI Program at the State of Alaska, Maternal, Child & Family Health, (907) 269-3400, or visit the program's web site at www.hss.state.ak/dph/mcfh

Alaska's Early Hearing Detection and Intervention Program Newborn Hearing Screening General Brochure includes basic information about newborn hearing screening, including information about what the screening is and why it is being done, and strives to represent all Alaskans by including photos of children from various ethnic backgrounds.

The brochure is intended for pregnant mothers, those women that have just given birth, and the general public. Ideal locations for delivery of the brochure include, but are not limited to, childbirth and prenatal classes, ob/gyns offices, mother baby units, birthing centers.

Alaska's Early Hearing Detection and Intervention Program "What's the Next Step?" Second Informational Brochure includes information about who parents should contact in their community for further testing regarding a suspected hearing loss and why this is important in terms of reducing or minimizing developmental delays. "*What's the Next Step?*" is intended for parents of newborns who refer, or do not pass, the hearing screening in the nursery, and will be available for nurses, audiologists and other health care providers who work with children and hearing loss, to deliver to parents as necessary.

Alaska's Early Hearing Detection and Intervention Program Basic Information for Parents of Children with Hearing Loss was developed in a format easy for parents to read and locate information about what should be done next after a hearing loss is detected, who to contact, why the hearing loss may have occurred, and other basics regarding how best for parents to help an infant/child with hearing loss. The brochure is a condensed version of the parent resource manual and may prove more appropriate for families to receive immediately following a confirmed hearing loss diagnosis, rather than the manual. This brochure will be available for nurses, audiologists, and other health care providers who work with children and hearing loss, to deliver to parents as needed.

Alaska's Early Hearing Detection and Intervention Program A Parent Resource Manual is in a format easy for parents to read and divided into sections to make it simple to locate information such as introductions, frequently asked questions, checklists, resources, and terminology. Included in the manual are topics including information regarding hearing loss, tests, early intervention, advocacy, communication options, and educational options.

For Healthcare Providers

The following resource was developed by Alaska's Early Hearing Detection & Intervention Program for Alaskan Healthcare Providers and is available for distribution.

Alaska's Early Hearing Detection and Intervention Program A Healthcare Provider's Guide is a concise booklet outlining information beginning with a child's primary healthcare provider's role as the medical home in the diagnosis of hearing loss. It serves as a referral guide for healthcare providers when presented with childhood hearing loss, including contact information of specialists for further testing and early intervention enrollment.

Alaska's Early Hearing Detection and Intervention Program Hospital Orientation Manual comprehensively details steps necessary to implementing successful newborn hearing screening programs in birthing facilities. This particular manual was developed for Alaskan communities specifically accounting for additional challenges placed on rural locations. The manual is available for birthing facilities interested in implementing newborn hearing screening programs in their communities.